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Acquired tubercular bronchoesophageal fistula in a hemophiliac child

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ABSTRACT

Acquired bronchoesophageal fistula in children is usually a late diagnosis, due to the rarity of the condition. The diagnosis was further complicated by presence of multiple co-morbid conditions. We would like to emphasize the importance of tackling the co-morbid factors strategically along with the surgical aspects in this case report.

Congenital broncho-oesophageal fistulae are commonly associated with oesophageal atresia. Acquired fistulae are rare in children. We present a case of an acquired Broncho-oesophageal fistula secondary to pulmonary tuberculosis in a known hemophiliac child with severe malnutrition. This made it a treacherous path requiring due diligence, caution and restrain to ensure an optimal outcome.

1. Case summary

A 7 yr cachectic boy, a known case of haemophilia had recurrent episodes of bronchopneumonia. He presented with frequent episodes of hacking, unrelenting cough and vomiting after ingestion of food. These episodes were aggravated with liquids than solid meals. Patient had bouts of hemoptysis and had lost 2 kg of weight in the preceding month. A dye study and HRCT (Figs. 1 and 2) demonstrated a fistula between the esophagus and the right lower bronchus along with consolidation of the right lower lobe. The challenging aspects were the correction of malnutrition, tuberculoses and control of haemophilia. Tuberculosis was confirmed after investigation and anti tuberculosis therapy was started. Patient was started on high protein diet through Ryle's tube to ensure optimal nutritional intake. Hematology opinion was taken for perioperative management of haemophilia. The patient was negative for factor VIII inhibitors and his factor activity was less than 1% indicating a more severe form of the disease. After adequate preoperative optimization, the fistula was defined and ligated (Fig. 3) with oesophageal repair over a Freka's tube by thoracotomy approach. A post-operative dye study performed on day 7, demonstrated a small leak which was managed conservatively. A repeat dye study 3weeks later demonstrated no leak (Fig. 4). Patient showed rapid weight gain after surgery and is doing well on follow up.

2. Discussion

Broncho-oesophageal fistula (BEF) is an abnormal communication between the bronchus and the esophagus causing recurrent respiratory tract infections. Congenital BEF is often seen in children. Acquired variety is generally seen in adults with malignancy, trauma, interventions and granulomatous conditions being the common causative factors [1]. In acquired fistulae, malignancy is a common cause in adults while granulomatous diseases like tuberculosis is commoner in children, especially from developing nations [2].

Tuberculosis leads to an immunocompromised state in the child, which hinders the healing process. Inflammation in and around these enlarged lymph nodes leads to the involvement of neighboring structures, particularly the esophagus and the trachea near its bifurcation, resulting in perioesophagitis and peritracheitis. It can form a traction diverticulum or if lymph node caseation and necrosis with abscess formation occurs then it can rupture and form a fistula. Less than 30 patients with tuberculous

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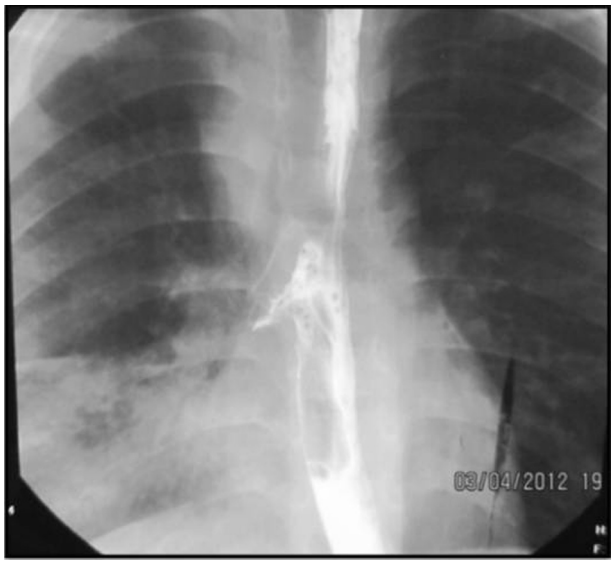

Fig. 1. Preoperative dye study with fistula.

broncho-oesophageal fistulae have been described in literature till date [2–4]. In addition, correction of malnutrition becomes a primary step in management.

The present case became more intriguing with addition of another co-morbid condition of haemophilia. Haemophilia is a rare disorder which is complex to diagnose and manage. It is an X linked recessive disease caused by deficiency of Factor VIII (haemophilia A) or deficiency of Factor IX (Haemophilia B). Perioperative management depends on level of factor activity or presence of inhibitors. Factor activity of less than 1%, causes spontaneous bleeding in form of haemarthroses, hemoptysis etc [5]. The optimal activity level for surgery should be between 60 and 80 IU/dl. This is achieved by transfusion of factor concentrates. In emergencies or unavailability of factor concentrates, fresh frozen plasma or cryoprecipitates can be transfused.

Surgical approach to an acquired BEF is the treatment of choice. Elective thoracotomy for excision and ligation of fistula and removal of diseased lung if any is preferred with high success rate. Recently conservative approach in the form of antituberculous or antiretroviral therapy, intercostal drainage and nutritional management has been published with favorable outcomes [6,7]. Endoscopic application of

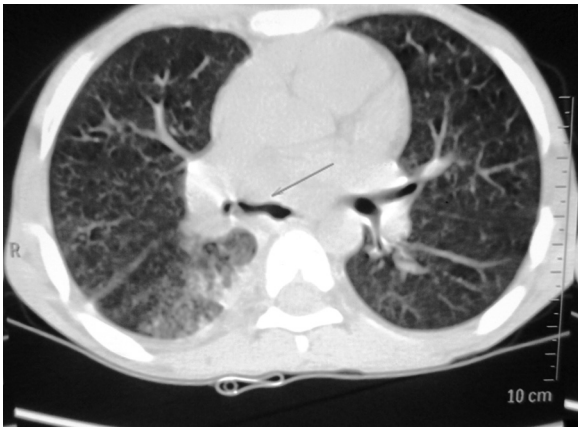

Fig. 2. HRCT demonstrating fistulous tract.

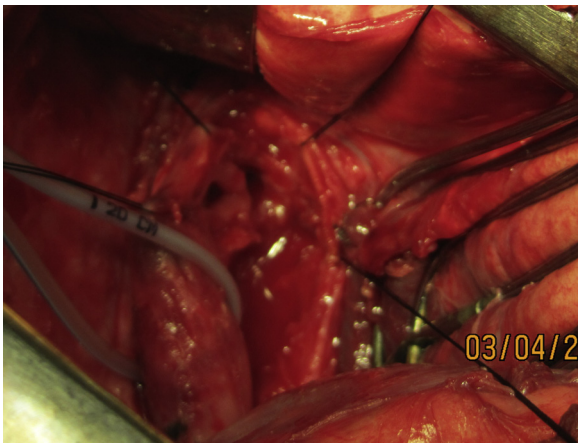

Fig. 3. Intraoperative photo.

silver nitrate, acetic acid had been tried earlier with high failure rate [8,9]. Endoscopic application of fibrin glue i.e. n-butyl 2 cyanoacrylate, coils or endoclip gives marginally better results but needs multiple applications. Attempts for a successful closure of fistula by vascular plugs or bypassing the fistulous tract with metallic stents have been published in literature. However a timely surgical intervention helps in complete eradication of the disease [10,11].

Diligence, patience and logical organized approach are important cornerstones to ensure optimal outcome for any patient. However their importance is highlighted in cases like these, which pose challenges in various aspects and demands a holistic approach. Although the preoperative preparations were exhaustive the final optimal outcome made it worthwhile.

Conflicts of interest

The authors declare no conflicts of interest.

Fig. 4. Post operative dye study on day 21: no leak.

References

[1] Baijal R, Ramegowda PH, Jain M, Gupta D, Shah N, Kulkarni S. Clinical profile and management of tuberculous bronchoesophageal fistula. J Dig Endosc 2013;4:103–6.

[2] Pecora DV. Tuberculous fistula of the esophagus. J Thorac Surg 1958;36:53.

[3] Nanaware SK, Gothi D, Joshi JM. Tuberculous bronchoesophageal fistula managed conservatively. Lung India 2005;22:65–7.

[4] Patel S, Abraham VJ, Mathur RM, Devgarha S, Yadav A. Acquired spontaneous bronchoesophageal fistula in an adult, Egypt. J Chest Dis Tuberc 2014.

[5] Ghosh K, Shetty S, Sahu D. Haemophilia care in India: innovations and integrations by various chapters of haemophilia federation of India (HFI). Haemophilia 2010;16:61–5.

[6] Raghu G, Dillard D. Esophagobronchial fistula and mediastinal tuberculosis. Ann Thorac Surg 1990;50:647–9.

[7] Shah CP, Yeolekar ME, Pardiwala FK. Acquired tracheooesophagealfistula. J Postgrad Med 1994;40:83–4.

[8] Sastry RA, Sanjeeva Rao K. Broncho-esophageal fistula: successful surgical repair after failed esophageal stent. J Med Sci Res 2014;2(3):153–7.

[9] Cossentino MJ, Ormseth EJ, Tavaf-Motamen H, Cheney CP. Congenital bronchoesophageal fistula in the adult: a case report. Am J Gastroenterol 2000; 95(8):2116–8.

[10] Rizzi A, Rocco G, Robustellini M, Rossi G, Della Pona C, Massera F. Results of surgical management of tuberculosis: experience in 206 patients undergoing operation. Ann Thorac Surg 1995;59:896–900.

[11] Takeda S, Maeda H, Hayakawa M, Sawabata N, Maekura R. Current surgical intervention for pulmonary tuberculosis. Ann Thorac Surg 2005;79:959–63.